CHURCH (A)

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Athetosis, with Clinical Cases

Church

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ATHETOSIS, WITH CLINICAL CASES.

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The condition first well clinically described by Hammond in 1871 and by him denominated Athetosis, is sufficiently familiar to neurologists, but in general practice is considered a very rare and curious anomaly, the significance of which is completely overlooked or misunderstood. Recently a number of pronounced and some rare instances of this form of diseased mobility have fallen under observation, and in every instance men of large experience in general surgical, and even orthopedic practice had not observed or had misinterpreted the phenomenon. That it is moderately rare is well shown by Osler's tabulation of 151 cases of cerebral palsies in children, in which it was noted eleven times, and is very properly differentiated from post-apoplectic chorea and tremor.

By European writers, as a rule, all excesses of motility following or associated with cerebral lesions are indifferently called choreic or spasmodic, but a careful or even superficial examination of a well-marked case of athetosis shows that in many respects there is no similarity whatever in the grotesque amoeboid activity of the athetoid digits to the gesticulatory movements of chorea or the implied rigidity of spasm. This distinction is to be insisted upon because, as is to be indicated later, athetosis is a localizing sign of presumptive value, and consequently of surgical importance, and always, probably, except in exceedingly rare hysterical instances, that should be readily detected, an indication of serious

organic brain disease. However, there may be an association of athetosis and chorea in the same case, both due to the same ultimate cause, or chorea of the ordinary type may occur in an individual presenting athetosis from birth, as I have once noted, without any material modification of either condition, or some rigidity may be present.

In Hammond's first case the symptom, for it can be called nothing more, appeared in an adult after a so-called "epileptic paroxysm" and was attended by great pain in the affected hand and foot, which were the seat of constant "complex involuntary movements," and these could not be controlled in any degree by the will, but persisted even during sleep. In the very large majority of instances, however, the condition dates from birth or shortly thereafter, is unattended by any impairment of sensation other than is common to the entire member or side of the body, is not marked by pain, and subsides during sleep or prolonged rest with support of the affected muscle groups and their congeners, and is intensified by emotion and most notably by voluntary effort to use the parts.

A somewhat full description of a case will afford an opportunity to discuss the various features of the condition consecutively and will avoid repetition.

Case I.—Johny B., aged 4 years (see cut 1), the child of healthy Irish parents, has presented peculiar movements of the right hand ever since birth. The mother was a primipara and the labor is described as extremely tedious, protracted and difficult, and finally terminated by the application of forceps to the head. The medical attendant is an especially skillful obstetrician and surgeon, and there is no doubt that every care and precaution was taken. A scalp wound over the right frontal eminence was caused and another over the left occipital region, both being considerable in extent and probably the former was attended with fracture as depression and adhesions are now present. He has never had any serious illness or convulsions and physic-

ally has been in excellent health. He has been slightly backward about talking and possibly is not as keenly intelligent as might be reasonably expected, but on the other hand is a bright, cheerful, active little fellow. From overaction of the right calf muscles he has always had difficulty in walking and there was a well-marked tendency to equine varus of the foot without bony deformity, a condition readily corrected by a plaster bandage, after wearing which

Cut 1.—From a Photograph.

for a few weeks the foot remained in good condition and is now, a year later, used with but a slight trace of difficulty. The hand is the seat of athetoid movements of a slow tentacle-like character, the position of the hand upon the wrist, and the fingers upon the hand, and in relation to one another, changing more or less continuously, and in a way suggestive of no volitional intention. If he attempts to grasp any object with this hand or to hold it still,

the athetoid activity is immediately increased and the fingers are, especially in prehensal attempts, widely and divergently extended, particularly at the metacarpo-phalangeal joints which, like those of the phalanges, are capable of a much wider range of extension than normal or than those of his left hand. This is most marked in the index which is capable of nearly as much movement in the direction of extension as in that of flexion. Finally when a small object is grasped it is with the palm, and the fingers take almost no part in the retention, which is uncertain at best. In this effort the entire member participates by being held rigidly at the shoulder and elbow and the over-activity of the right side of the face becomes apparent, (see cut 1,) showing, with the difficulty in the gait and foot, the hemiplegic distribution of the trouble. The facial over-activity is particularly marked in all expressions of emotion, suggesting a weakness of the left side which, however, is only apparent. This state of over-action on the right side is analagous to the late over-action in other conditions of a hemiplegic character and has been found in all the cases that I have been able to examine.

The muscles of the forearm and hand are firm, well developed, and when their incoördinate action permits, a test shows a fair degree of strength. The forearm measures slightly more than on the unaffected side. The over-development or full development of muscles subjected to athetoid activity is always noticeable and atrophy is never present.

In this case sensation in all its factors seems perfect, though the age of the patient precludes the most delicate examination. It is certain that touch, temperature, pain and pressure are fairly well distinguished, as is also the sense of position, for with closed eyes he will represent with the unaffected hand the various distortions assumed by the athetoid member and locate its position. The reflexes throughout the right side are in slight excess of the left, no

ankle clonus, nystagmus, inequality of pupils, variation to electrical control or impairment of trophic function in epidermal structures are present.

At the first glance one would be inclined to attribute to the use of forceps and the following local injury a causal rôle, though the anatomical reason for it would be hard to bring forward. But there are other factors of equal or more importance. The prolonged labor, which in itself probably is sufficient to account for the trouble, must not be forgotten. Indeed, Osler very reasonably urges that the association of cerebral palsies, with protracted labor, is an urgent indication for forceps in its prevention, and many pædiatrists can be quoted to show that the use of forceps almost never in itself produces serious injury to the brain even in the cases where external appearances would lead one to apprehend such an accident. It was also incidentally discovered that the child's uncle on the mother's side presents a most typical case of athetosis. With him the left side is affected, the hand being in constant motion, even during sleep. In this case no use of forceps or difficult labor had taken place. I do not know that such relationship has been heretofore noted, but am disposed to question whether there may not be a family tendency to such accidents aside from the inheritance of narrow pelves in the females, and other causes of tedious labor, just as a tendency to apoplexy late in life is not infrequently observed to be a matter of clearly marked heredity.

The next case is presented because of the symmetrical and universal distribution of the athetosis, which is of a pure type. Double athetosis is not much more rare than the monoplegic or hemiplegic variety, but usually is marked by spasticity, inability to walk, great mental enfeeblement or idiocy, and is not infrequently denominated spastic paraplegia, the rigidity and increased myotatic irritability being the most pronounced features.

Case II.—Alvina W., single, female, age 30 years. Some what lacking in mental development, but not by any means idiotic. She states that at the time of her birth the midwife made some mistake, but what it was she can not tell nor can any particulars be learned and there is no external indication of injury to the head or scalp. She has never suffered from any serious physical illness and aside from her athetoid condition is in fair health and strength, of a cheerful, bright disposition, able to get about readily, to dress herself, make her bed and lend a little assistance to those about her in the County Infirmary. When sitting at rest nothing very noticeable is seen, but upon the slightest attempt to speak or to move all her extremities and particularly the fingers and toes, and her face, head and eyes are started into peculiar uncertain, rather haphazard contortions, which under effort or emotion of any sort, extend to the upper parts of the limbs and the trunk. The fingers become widely spead out extended and flexed, either in combination or separately, yet she is able in a way to direct them as in buttoning her dress, which is accomplished laboriously and only after much time. The effort to do this will cause her to perspire freely and show fatigue. The toes present the same athetoid movements in a very high degree. Owing to the constant muscular activity and the tendency of the toes to spread laterally and work individually, the form of the foot is considerably changed having a triangular outline with almost as much distance between the great and little toes as from the fifth toe to the heel. The implication of the face is also symmetrical, exaggerating and caricaturing every facial expression. In the cut (2) taken from a "snap shot", she is trying to sit quietly, but was pleased at having her picture taken and the resulting smile becomes the distortion indicated. It also shows the overactivity of the muscles of the neck and the movements of the fingers. The constant action has resulted in an unusual development of the musculature of the entire body so that

she presents a rather masculine figure and doubtless the skeleton itself has been correspondingly modified. Even the muscles of respiration and vocalization are implicated. Her speech is halting, stammering, spluttering, explosive and modified in every way, resembling no other speech defect with which I am acquainted, presenting to the ear the disordered muscular action which in the extremities is patent to the eye. There is also a disorderly nystagmus in which

Cut 2.—From a Photograph.

the eyes move laterally, obliquely and vertically independent of each other and which, with all the other conditions noted, ceases during rest and is correspondingly emphasized by emotion. The special senses are not notably impaired in any way and general sensation is intact in all its forms. Response to electricity is normal. All reflexes are exaggerated. There is no ankle clonus or loss of sphinteric control.

Case III.—Thos. C., aged 13, of strong, vigorous, healthy, English parents, born without difficulty, head presenting, labor lasting about two hours. Peculiar movements of all extremities have been present since birth. Has never had any serious illness or convulsions. The boy is somewhat backward mentally, principally from lack of instruction, most of his life having been passed in the County Infirmary. He reads some, answers questions intelligently and is quick and observing. His physical health is fairly good, but he is not large for his age. With the exception of the

Cut 3.—From a Photograph.

face and neck athetoid movements are present in the entire body, most pronounced, contrary to the usual rule, in the lower extremities, which, during the waking hours, are in nearly constant contortions from the pelvis down, and so vigorous are they, that a strong pair of shoes will be rubbed to pieces in the course of a few weeks. Though he is unable to walk he manages to crawl around on elbows and knees in a wriggling, uncertain, but rather rapid way, causing,

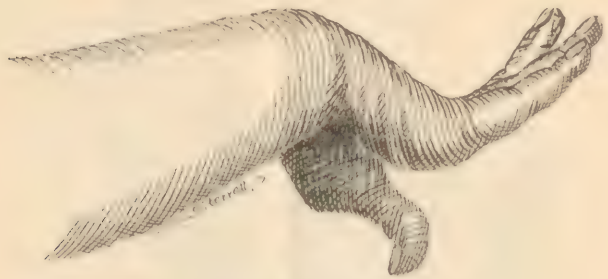
with the constant friction incident to the athetosis, numerous callosities about the joints. During sleep all movements cease and, when quietly resting in his chair or bed, the trunk and upper extremities are free from movement, but immediately manifest it upon voluntary effort or emotion. Prehension is practically impossible. (See cuts 3 and 4.)

There is slight voluntary power to direct movements of the arms and legs, but their course is subject to wild excursions, reminding one of the incoördination of advanced ataxia and attended with an aggravation of the athetoid

Cut 4.--From a Photograph.

features. The peculiar and characteristic condition in the feet is shown in cut 4. Speech is also modified as in the preceding case. Special senses, general sensation, control of sphincters and trophic conditions are all normal. There is an exaggeration of all reflexes; the muscles are fairly developed and firm; there is a tendency to "cross-leg" and the over-action of the calf muscles has produced pronounced deformity at the ankle joint, which can, however, be very nearly reduced by manual manipulation.

With such disorderly movements and total irregularity of muscular action it is difficult to generalize, but in these and other cases there is observed a preponderating action, in the upper extremities, of flexion at the wrist and extension and divergence of the digits (see cut 5) and, in the lower extremities, the homologous groups of muscles are similarly most affected, producing equine position of the foot with extension and spreading of the toes. The ankle joint is the only one that I have found notably deformed though the increased range of motion in the smaller articulations has been already noted.



Cut 5.—From a Photograph.

The lesion in this condition according to Hammond, who has collected the reports of thirteen autopsies, invariably involves the optic thalamus and the lenticular nucleus of the striate body, one or both, or the motor cortex corresponding to the affected muscles. In none of these reported cases was the motor tract destroyed and consequently the local condition must be of an "irritation" character, a sort of discharging lesion constantly operating.

Where the lesion in the basal ganglia infringes upon the capsule it determines motor and sensory symptoms with anatomical exactness, but in the cases so far reported the interference with the capsule has usually spared the sensory portion. In double athetosis it is reasonable to suppose that the lesion must be cortical, as it is difficult to imagine symmetrical damage at the base, while the vulnerability of

the cortex and its liability to inflammatory processes which are capable of producing permanent effects, lays it liable to widely distributed lesions not rarely symmetrical. Its appearance, too, in cases marked by general cerebral atrophy and wide spread sclerosis is indicative of a cortical distribution in the symmetrical form of the affection. At any rate one is now justified in locating the disturbance in every case of athetosis in one or the other position and cognate conditions, must determine, which it probably is while its nature may be fairly inferred from the clinical aspects of the case.

Regarding treatment there is medically very little that can be done. Hyoscyamus, cannabis indica, gelseminum and similar sedatives will reduce and temporarily modify the movements, but disturb the general health and their withdrawal is promptly followed by a full recurrence of the original state. Surgical measures have been advocated by some, and Horsley goes to the extreme of recommending, that every case of athetosis should be operated upon and the related cortical centres removed. He certainly did not have the double form in mind, when giving this advice. Where, however the athetosis as in Case I. is practically limited to a few muscles, the proposition may be considered. Such decortication of course produces paralysis in the member which may or may not be preferable, and few patients would willingly accept the exchange. If the hope and probability of a return of voluntary power in the hand should be well founded, from the vicarious control of associated or symmetrical cerebral centres a great gain and practical cure would result. To attack the basal lesion is of course impossible. The procedure of Hammond in stretching the peripheral nerves though it gave some relief for a few months each time in his first case, as reported by him, can not be reasonably recommended.

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